A MODIFIED REPAIR TECHNIQUE FOR TRICUSPID INCOMPETENCE IN EBSTEIN'S ANOMALY

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Objective: **A modified technique for tricuspid valve repair in Ebstein's anomaly restructures the valve mechanism at the level of the true tricuspid anulus by using the most mobile leaflet for valve closure without plication of the atrialized chamber. Midterm results of this therapeutic approach for patients with Ebstein's anomaly and tricuspid valve incompetence are reported.** ***Methods:*** **Between October 1988 and April 1997, the incompetent tricuspid valve was repaired with our technique in 19 patients (12 female, 7 male; 2 to 54 years, mean 21 years). The indication for operation was congestive heart failure of various degrees in all patients. Tricuspid incompetence was grade II in two patients, grade III in 14, and grade IV in three. Associated congenital malformations were simultaneously repaired (interatrial communication in 18, ventricular septal defect in two, pulmonary stenosis in two, mitral valve prolapse in one). Follow-up ranged between 10 and 103 months (median 28 months) and was complete for all patients.** ***Results:*** **There were no operative deaths. One patient with active endocarditis and pulmonary abscess died 2 months after the operation of recurrent sepsis; there were no late deaths. During follow-up, New York Heart Association functional class improved from 2.8 before the operation to 1.9 without recurrent cyanosis, and tricuspid incompetence decreased from a mean grade of 3.1 to one of 0.9, without any echocardiographic deterioration of the tricuspid valve function or right ventricular dilation.** ***Conclusions:*** **Our technique allows tricuspid valve repair in patients with Ebstein's anomaly, even in cases usually reserved for primary valve replacement, without late functional deterioration. (J Thorac Cardiovasc Surg 1998;115:857-68)**

Ebstein's anomaly, a complex congenital defect of the tricuspid valve and right ventricle originally described in 1866 by German pathologist Wilhelm Ebstein,[1] accounts for approximately 0.3% of all congenital heart defects seen in a large pediatric center.[2] At our institution, this type of defect accounts for 1.2% of all cardiac surgical procedures for congenital heart disease. Its most prominent pathologic characteristic is that the septal or posterior leaflets of the tricuspid valve are variably deformed and displaced downward into the right ventricle, without normal attachment to the tricuspid valve anulus. The anterior leaflet is usually enlarged and sail-like but in the normal position. The displacement of the valve toward the apex of the right ventricular cavity divides the right ventricle into "atrialized" and "true" ventricular portions.[3, 4] The anulus of the tricuspid valve is almost always dilated, and the valve itself is usually incompetent. An interatrial communication, usually a patent foramen ovale or atrial septal defect (ASD), is a frequent finding, accompanied by right-to-left shunt and systemic arterial desaturation and cyanosis.[5]

Since the first surgical attempts,[6-9] operative treatment has been disputed regarding indications and suitable techniques (valve replacement or repair).[3, 10-16] We introduced our technique in 1988; since 1994, it has been our standard surgical repair technique for patients with Ebstein's anomaly and

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Table I. *Demographic characteristics, symptoms, and findings in 19 patients with Ebstein's anomaly undergoing tricuspid valve repair (Deutsches Herzzentrum Berlin, October 1998 through April 1997)*

No.	*Age (yr)*	*Sex*	*Date of operation*	*Follow-up (mo)*	*NYHA class (preoperative/ postoperative)*	*TV incompetence (preoperative/ postoperative)*	*SaO_2 (% preoperative/ postoperative)*	*IV m/sec*	*Rhythm (preoperative/ postoperative)*
1	14	F	10/1988	103	3/2	2/0-1	92/99	0.6	AF/AV third degree
2	17	F	8/1992	56	3/2	3/1-2	97/98	1.2	SR/SR
3	12	F	1/1993	51	3/2	3/1-2	97/98	0.7	WPW/WPW
4	5	M	11/1993	42	2/1	2/0-1	97/98	0.6	SR/SR
5	16	M	3/1994	37	2/2	3/0-1	98/98	0.7	SR/SR
6	34	M	3/1994	37	2/2	3/0-1	100/100	0.6	AF/SR
7	12	F	4/1994	36	4/2	4/1-2	84/100	0.6	SR/SR
8	49	F	12/1994	29	4/3	4/1-2	76/98	0.7	SVT/SR
9	10	F	1/1995	27	2/1	3/0-1	98/98	0.8	SR/SR
10	10	M	4/1994	24	4/3	3/1-2	77/98	0.7	SR/SR
11	33	F	5/1995	23	4/2	4/1-2	83/96	1.3	AV first degree/ AV second to third degree
12	20	M	10/1995	19	3/2	3/0-1	93/99	0.9	SVT/SR
13	54	F	12/1995	2 (died)	3/-	3/0-1	89/98	-	AF/SR
14	14	F	1/1996	15	3/2	3/1-2	100/100	1.3	SR/SR
15	14	M	2/1996	14	3/2	3/0-1	78/96	0.9	SVT/SR
16	5	M	4/1996	12	3/1	3/0-1	93/100	0.6	SR/SR
17	16	F	5/1996	11	2/1	3/0-1	98/98	1.0	SR/SR
18	53	F	6/1996	10	4/2	3/1-2	91/95	0.5	SVT/SR
19	2	F	6/1996	10	3/2	3/0-1	90/95	0.7	SR/SR

NYHA, New York Heart Association; *TV,* tricuspid valve; *Sao_2,* arterial oxygen saturation; *IV,* inflow velocity; *AF,* atrial fibrillation; *AV,* atrioventricular block; *SR,* sinus rhythm; *WPW,* Wolff-Parkinson-White syndrome; *SVT,* supraventricular tachyarrhythmia; m/sec = meter per second.

tricuspid valve incompetence. Our technique restructures the valve mechanism at the level of the true tricuspid anulus by using the most mobile leaflet for valve closure without plication of the atrialized chamber. This concept was based on the assumption that it may not be necessary to exclude the atrialized chamber, and that it is desirable to restructure the valve mechanism at the level of the true tricuspid anulus by using the most mobile leaflet for valve closure. This study evaluated our midterm results with this therapeutic approach.

Patients and methods

Patients and their preoperative characteristics. Nineteen patients (12 female, 7 male; age 2 to 54 years, mean 21 years) with Ebstein's anomaly and tricuspid valve incompetence underwent our modified repair technique between October 1988 and April 1997 (Table I). Since March 1994, no patients other than the reported cases were operated on at our department, except for one patient who primarily underwent tricuspid valve replacement for Carpentier type D anatomy[13] where repair was deemed not feasible. The indication for operation was congestive heart failure of various degrees in all patients (Table I). One patient had systemic embolism and 11 had cyanosis with systemic oxygen saturation at rest of between 76% and 92%. All patients had undergone preoperative echocardiographic studies and right and left heart angiography. Diagnosis was established before the operation in all cases, except for that of a 51-year-old-woman in whom a device occlusion of the ASD attempted at the Cardiology Department was followed by dislocation of the device and right heart endocarditis with pulmonary septic embolism. The correct diagnosis of Ebstein's anomaly was made during the emergency operation.

Findings at operation and types of procedures. Findings are listed in Table II. Operations were carried out through median sternotomies with separate caval cannulation, total cardiopulmonary bypass, and mild systemic hypothermia of 30° C. Cardioplegic arrest was initially provided by aortic root infusion of crystalloid arrest solution (Cardioplegin), and myocardial protection was maintained by repeated infusion of cold hydroxyethylene starch solution. The right atrium was entered through an angulated oblique incision and the situs was examined. As is typical for this complex defect, the pathologic anatomy varied widely in all patients, and no cases were alike in every respect; however, the characteristic features of Ebstein's anomaly were easily found in every patient. The anatomic anulus of the tricuspid valve was grossly distended, and the septal leaflet was displaced in its origin toward the right ventricular cavity. The posterior and the anterior leaflets showed marked variety. The annular attachment of the posterior leaflet was displaced, giving rise to the formation of an atrialized chamber of greatly differing extent. This chamber occupied a moderate portion of the entire right ventricle (equivalent to Carpentier's type A anatomy[13]) in four patients, and the atrialized chamber took in the larger part of the right ventricle in 15 cases (Carpentier's type B and C anatomy[13]), leaving

Table II. *Intraoperative findings in 19 patients with Ebstein's anomaly undergoing tricuspid valve repair (Deutsches Herzzentrum Berlin, October 1988 through April 1997)*

No.	*Carpentier type*	*Leaflet findings*	*Type of reconstruction*	*Repair of associated anomalies*
1	C	Fissures AL	Double ostium	ASD II patch closure
2	A	Cleft AL	Cleft closure; posterior anuloraphy	ASD II patch closure
3	A	Typical findings	Posterior annulorrhaphy	ASD II patch closure; PS homograft
4	A	Typical findings	Posterior annulorrhaphy	ASD I patch closure
5	B	Typical findings	Bilateral annulorrhaphy	ASD II direct closure
6	B	Large PL; fissures AL	PL translocation; bilateral annulorrhaphy	ASD II direct closure, VSD patch
7	B	Clefts AL	Cleft closure, posterior annulorrhaphy	ASD II direct closure
8	B	Typical findings	Anterior annulorrhaphy	ASD II direct closure
9	A	Large PL	PL translocation, posterior annulorrhaphy	ASD II direct closure; Chiari network
10	C	Restrictive AL	AL translocation, bilateral annulorrhaphy	ASD II patch closure
11	C	Accessory antero-septal leaflet	Posterior annulorrhaphy	ASD II direct closure
12	B	Typical findings	Bilateral annulorrhaphy	ASD II direct closure
13	C	Typical findings, restrictive AL Active endocarditis, pulmonary abscess	Posterior annulorrhaphy	ASD II patch closure ASD device removed
14	B	Fissures AL	Posterior annulorrhaphy	ASD II patch closure
15	B	Typical findings	Posterior annulorrhaphy	ASD II patch closure
16	B	Typical findings	Posterior annulorrhaphy	ASD II patch closure
17	B	Typical findings	Posterior annulorrhaphy	
18	C	Typical findings	AL mobilization, posterior annulorrhaphy	ASD II patch closure
19	B	Typical findings	Posterior annulorrhaphy	ASD II direct closure; PS homograft

AL, Anterior leaflet; *PS,* pulmonary stenosis; *PL,* posterior leaflet; *VSD,* ventricular septal defect.

only a smaller trabeculized right ventricle toward the apex, below the anterior leaflet, and in the outflow tract. The anterior leaflet was largest in all cases; however, the posterior leaflet was quite substantial and mobile in eight cases and reached almost the same size as the anterior leaflet in four instances. The anterior leaflet itself showed clefts or deep fenestrations in four instances, and in one case there was a fourth, smaller, accessory leaflet with origin in the anteroseptal commissure. The anterior leaflet in five patients had localized or wide-spread chordal attachment to the right ventricular free wall, restricting leaflet mobility (Carpentier's type C anatomy,[13] Table II).

Operative technique. The technique is illustrated in Figs. 1 through 4. The basic principle of the valve reconstruction in this group of patients was to reduce the anatomic tricuspid ostium so that the most mobile leaflet, mostly the anterior leaflet or part of it or in some instances the posterior leaflet, may find an opposing structure for coaptation during systole. This concept was first applied in the case of a 41-year-old woman with type C anatomy in whom, after a widely fenestrated anterior leaflet was freed from adhesions with the right ventricular free wall, it was believed that no competence would be obtainable with the Hardy technique.[3] In this case the huge ostium was divided into halves by passing two strong mattress sutures from the middle of the anterior anulus to the middle of the atrialized septum just below the natural anulus. Through this approximation of the two opposite parts of the anulus, in each of the resulting two ostia the unfenestrated portion of the anterior leaflet could then coapt with the opposing septum. This repair has remained competent now for almost 9 years.

The next two patients (cases 2 and 3) in whom the modified technique was applied had been operated on previously. Although primary reconstructive results were good after tricuspid anulus shortening attempted by suture annuloplasty along the posterior and the anterior natural anulus similar to the De Vega annuloplasty, valve incompetence recurred during follow-up and the patients had to be operated on again at 19 and 24 months, respectively, with the modified technique. In both patients, who underwent reoperation in 1992 and 1993, respectively, the posterior half of the natural ostium was completely closed. Since then, tricuspid incompetence has remained minimal in both cases.

Since these initial experiences, the technique has been standardized according to the following concept. On careful inspection of all leaflet components, it was determined which leaflet part was the most mobile and substantial and also had sufficient free edge chordae. In two

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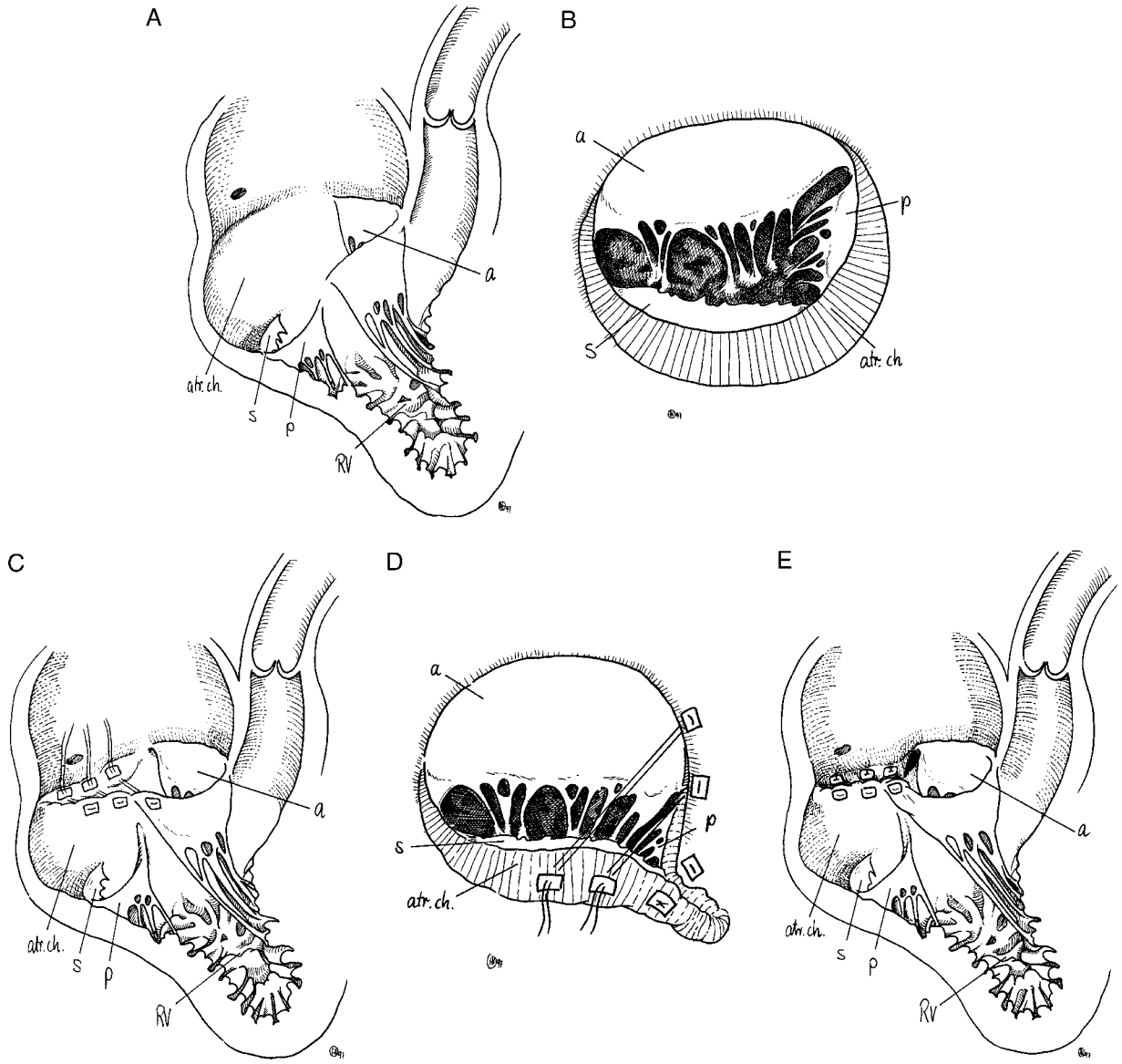

Fig. 1. Schematic presentation of a cross-sectional **(A, C, E)** and surgeon's views **(B, D)** of the intraoperative findings and the surgical technique applied in patients 2, 3, 4, 11, and 13 through 19. **A** and **B,** The pathologic finding corresponds to Ebstein's anomaly of Carpentier type B,[13] with the displaced septal *(s)* and posterior *(p)* leaflets toward the apex of the right ventricle *(RV).* Leaflet displacement creates an atrialized chamber *(atr. ch.)* below the anatomic tricuspid anulus. There is a large, mobile anterior leaflet *(a).* **C, D,** and **E,** The anterior part of the anterior leaflet was chosen for the valve-closing structure. A mattress suture of 3-0 polypropylene pledgeted with autologous pericardium is passed from the anterior leaflet anulus to the atrialized septum just below the natural tricuspid anulus. A row of these sutures is added toward the posterior anulus. These pledgeted stitches can also be placed through the septal anatomic anulus. After the sutures are placed they are tied, resulting in obliteration of the posterior half of the anatomic tricuspid orifice. Thus the anterior anulus is approximated to the septum. When valve competence is tested by filling the ventricular cavity with saline solution, the anterior part of the anterior leaflet now coapts with the atrialized septum. The "atrialized chamber" is now incorporated into contracting right ventricular cavity.

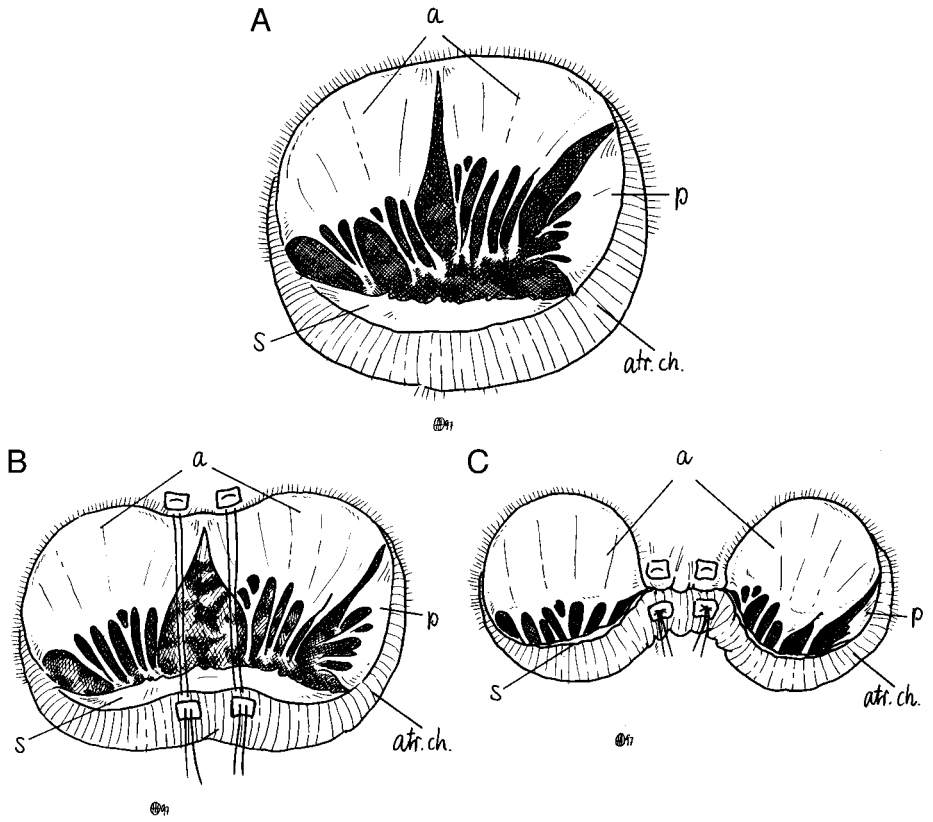

Fig. 2. A view of the tricuspid valve in case 1 from the surgeon's side. **A,** The anterior leaflet *(a)* is split by a cleft and shows deep fissures. There are some chordal adhesions of the leaflet to the right ventricular wall, which are dissected. **B,** Two 3-0 polypropylene mattress sutures supported with autologous pericardium pledgets are passed through the middle portions of the anterior anulus and the atrialized septum just below the septal anulus. **C,** When the sutures are tied, the anterior anulus is approximated to the septum, dividing the valve into two orifices and enabling valve closure by the solid part of the leaflets, which can now meet the atrialized septum and afford valve competence. *p,* Posterior leaflet; *s,* septal leaflet; *atr.ch.,* atrialized chamber.

cases of type C anatomy, mobility of a restricted anterior leaflet was enhanced by dissection of the free wall chordae. To ensure sufficient leaflet body, deep fissures or clefts of the anterior leaflet were closed by sutures in two patients. Once the most mobile part was chosen to be used as the main valve-closing leaflet, the remainder of the natural anulus was obliterated at the level of the natural anulus, either by sewing anulus to anulus or by using a large adjacent leaflet part to close the desired ostial portion. In no case was plication of the atrialized chamber attempted. In a large proportion of our patients (10 cases), the posterior half of the tricuspid ostium was closed directly. In two cases the posterior leaflet was sutured with its septal edge to the septum, in three cases most of the anterior part of the anterior leaflet was sewn to the septum, and in five instances the ostium was narrowed at both the anteroseptal commissure and the posterior anulus. In general, 3-0 polypropylene single mattress sutures supported by pledgets of autologous pericardium were used. The sutures on or along the septum were positioned into the muscular part just below the natural anulus, except for those along the posterior septum where sutures were run through the thin fibrous structure representing the natural anulus (Table II). The repair was evaluated by repeated filling of the ventricle with saline solution to make sure that the leaflet chosen as closing mechanism could in fact fulfill this task satisfactorily. The diameter of the residual valve ostium was measured and was never smaller than 23 to 25 mm. Once the heart was closed and beating, intraoperative transesophageal echocardiography was carried out to assess the valve and heart function.

Additional cardiac procedures. All but two patients had either secundum ASD (nine patients) or patent foramen ovale (eight patients), which were closed directly or with a patch of autologous pericardium in eight cases. One patient had no interatrial communication, and one had partial atrioventricular (AV) canal defect with a cleft in the anterior mitral leaflet. The cleft was closed by direct suture, and the primum ASD was closed with a pericardial

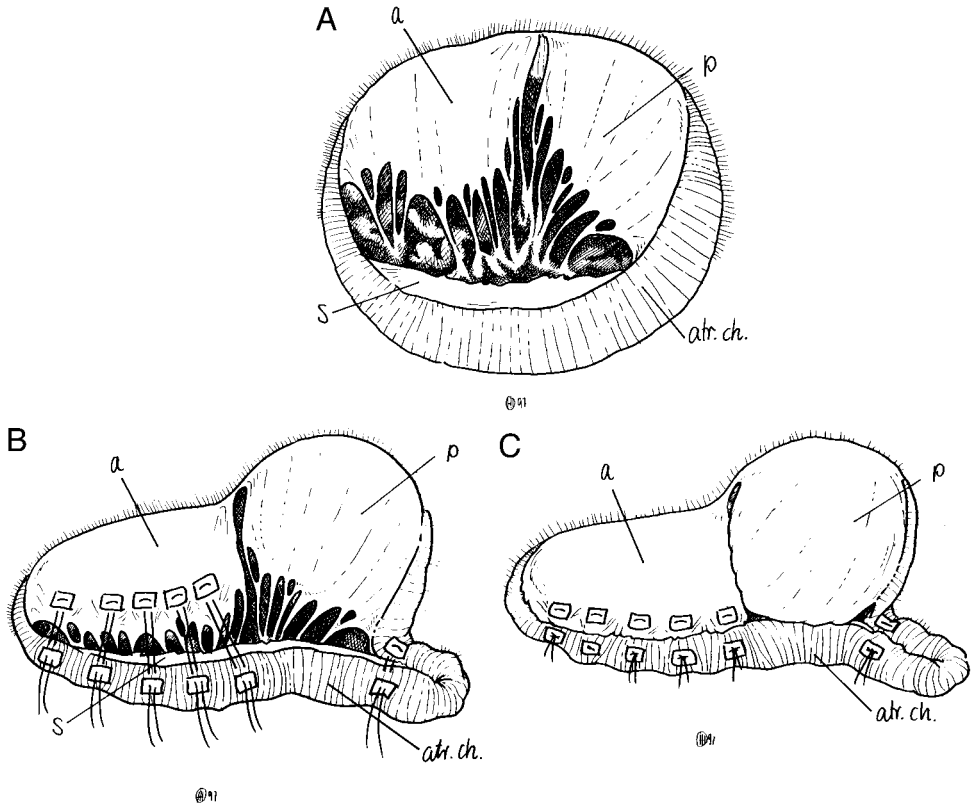

Fig. 3. Schematic presentation of the valve repair technique used in cases 5, 8, and 10. **A,** Anterior *(a)* and posterior *(p)* leaflets are almost equal in size. In case 10, there were additional chordal adhesions between the anterior leaflet and the right ventricular wall. The large posterior leaflet was selected for valve function. **B,** The anterior leaflet was translocated to the atrialized septum by a row of 3-0 polypropylene pledgeted sutures. Furthermore, the posteroseptal angle of the anatomic anulus was plicated for closer approximation of the anterior anulus to the septum. The anterior half of the orifice is now closed with the anterior leaflet, and the posterior leaflet moves freely. **C,** Testing of valve competence reveals appropriate valve closure achieved by good coaptation of the posterior leaflet with the septum. *s,* Septal leaflet.

patch. In a patient with a perimembranous ventricular septal defect situated just below the anteroseptal commissure, the ventricular septal defect was closed with a patch and the commissure was closed over a 2 cm length above the patch. One patient had an extensive Chiari network in the right atrium, which was resected. Two patients with high-grade valvular and subvalvular pulmonary stenosis received cryopreserved valved homograft conduits implanted between the right ventricular outflow tract and the main pulmonary artery. The patient who was operated on during the course of active endocarditis also showed mitral valve prolapse with moderate mitral incompetence, which was treated during the same operation by posterior anulus shortening with the Paneth annuloplasty.[17] The closure of the right atrial incision was accompanied by a substantial reduction in right atrial volume, which was considered necessary to reduce the size of the heart, which is frequently huge, and give the lung more room in the chest.

Follow-up. All patients have been prospectively followed up on a regular basis at least once a year. Clinical examination, echocardiographic examination, electrocardiography, and Holter monitoring were performed during the regular surveillance. Follow-up was complete in all cases and lasted between 10 and 103 months (median 28 months).

Results

Survival. All patients survived the operation, and there were no major perioperative or immediate postoperative complications. One patient (patient 13) who had been operated on during the course of septic endocarditis and pulmonary abscess initially recovered well; however, recurrent sepsis and mediastinitis developed and were treated by mediastinal irrigation and omentum translocation. Unfortunately, she died of sepsis 8 weeks after the initial

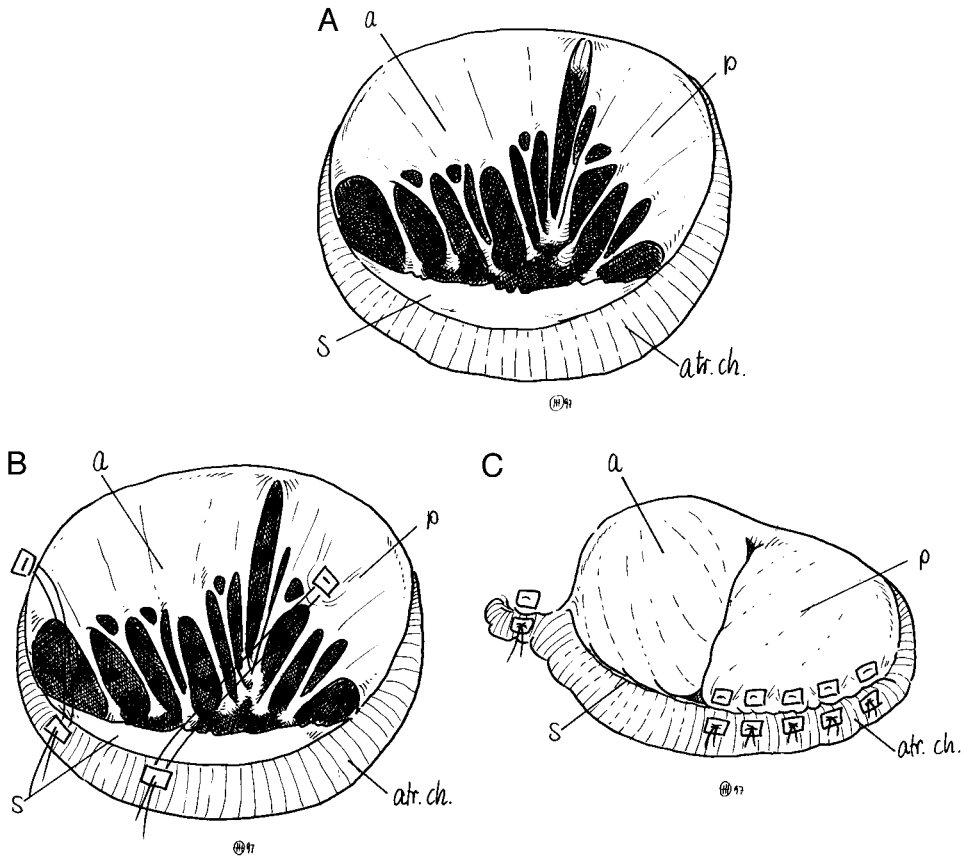

Fig. 4. Schematic drawings of pathologic findings and applied surgical technique in cases 6 and 9. **A,** The anterior *(a)* and the posterior *(p)* leaflets are of almost equal sizes. The anterior leaflet is mobile, but too small to close the orifice by itself. **B,** The posterior leaflet is transposed with its septal free edge to the atrialized septum below the anatomic anulus. In addition, the anteroseptal commissure is plicated by a pledgeted suture. **C,** On valve closure, the anterior leaflet plays the main closing part, which coapts with the fixed posterior leaflet and the atrialized septum. *s,* Septal leaflet; *atr.ch.,* atrialized chamber.

procedure. All other patients were alive as of the end of April 1997.

Postoperative rhythm. Two patients had permanent third-degree AV block that necessitated pacemaker implantation. Two more patients had transient AV block that disappeared shortly after the operation. One patient with known Wolff-Parkinson-White syndrome underwent successful catheter ablation of her abnormal pathway after the corrective operation (patient 3). Of five patients who had supraventricular tachyarrhythmic disturbances, one had postoperative third-degree AV block (patient 1) and the tachyarrhythmia disappeared and normal sinus rhythm has since persisted in four patients.

Functional capacities (New York Heart Association functional class). All long-term surviving patients had improvement of their functional capacity, except for two patients (patients 5 and 6) who had had only mild preoperative symptoms and stayed in the same New York Heart Association class II after the operation, although their preoperative tricuspid valve incompetence was significantly reduced. Improvement was achieved by one class in 12 cases and by two classes in four (Table I). New York Heart Association functional class improved from the preoperative mean of 2.8 to the postoperative value of 1.9.

Cyanosis. Only one patient had no intracardiac communication; all the others displayed cyanosis at rest or during exercise. Arterial oxygen saturation at rest was normal (>95%) in eight patients; cyanosis was present at rest in 11, with arterial oxygen saturation values between 76% and 92% (mean 86%). After the operation, all patients had normal oxygen saturation values (Table I).

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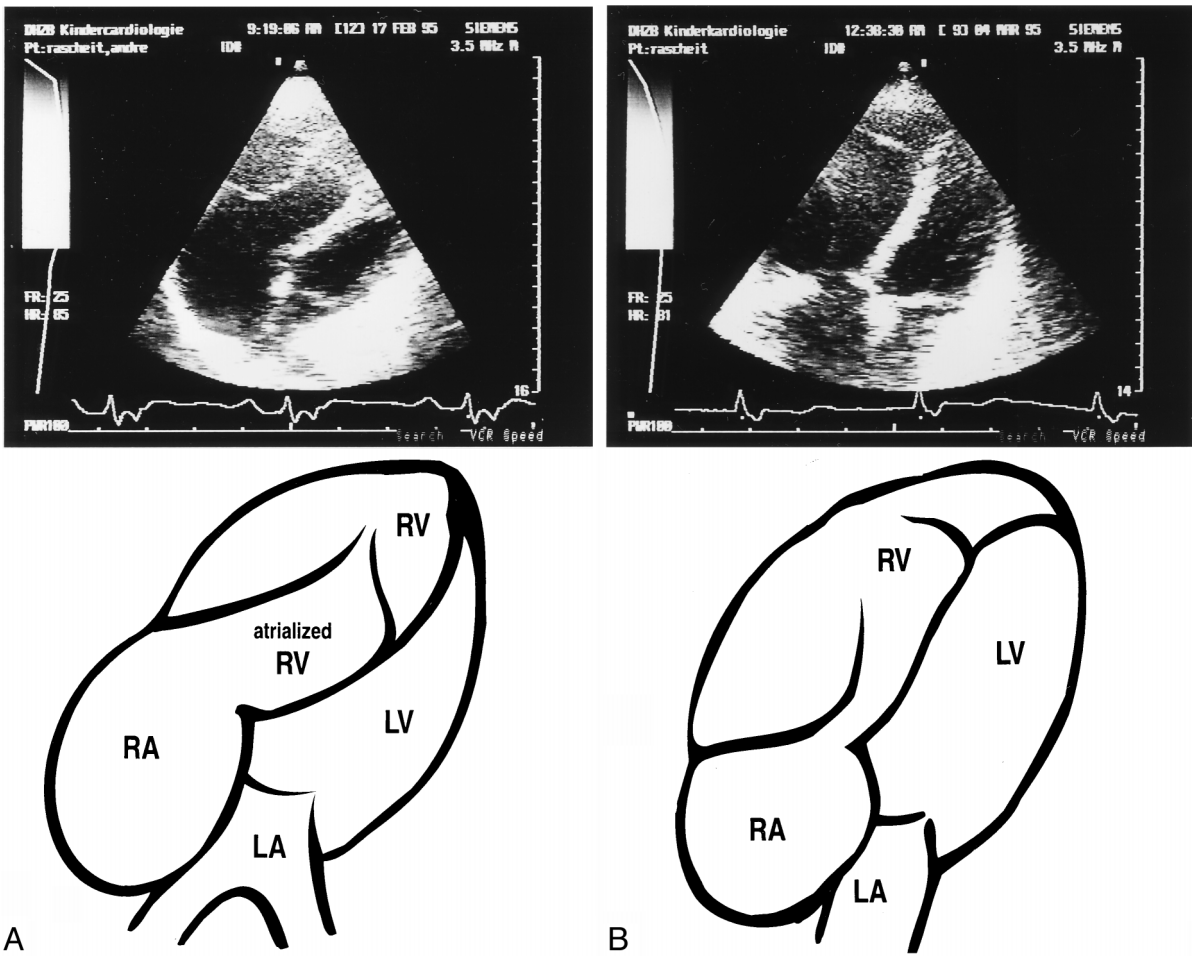

Fig. 5. Four-chamber echocardiographic views and schematic drawings of case 15. **A,** Typical preoperative echocardiographic findings of Ebstein's anomaly with a large mobile anterior leaflet, a sizeable atrialized chamber, and the septal leaflet displaced toward the right ventricular *(RV)* apex. **B,** Postoperative findings 1 year after tricuspid valve repair by posterior annulorrhaphy. Note the right ventricle has gained size and an almost normal shape by unification of the atrialized chamber with the true right ventricular cavity. The echo-dense structure posterior to the newly created, much smaller tricuspid orifice corresponds to the closed ostial part. *LV,* Left ventricle; *RA,* right atrium; *LA,* left atrium.

Echocardiographic assessment of tricuspid valve function before and after the operation. Tricuspid incompetence, the most dominant clinical finding other than cyanosis, was found in all patients. Tricuspid incompetence was graded II in two patients, III in 14, and IV in three. After the operation tricuspid incompetence regressed significantly in all cases, and there was no instance of tricuspid incompetence graded higher than II. A small jet was seen passing along the septum in all patients; this, however, was estimated to be minimal (0 to I) in 11 cases and I to II in eight.

Because the amount of tricuspid orifice reduction in some of the individual repair techniques was quite substantial, including obliteration of more than half of the originally huge anatomic anulus, a concern was raised regarding whether tricuspid stenosis might ensue. Echocardiographic follow-up studies revealed inflow velocity between 0.5 and 1.3 m/sec in most patients, and velocity was higher than 1.0 m/sec in only three, without clinical relevance (Table I). This corresponded well with the intraoperative evaluation of the residual valve orifice, which was always larger

Table III. *Different surgical techniques for tricuspid valve repair in Ebstein's anomaly*

Technique	*Plication of atrialized chamber*	*Closing leaflet*	*Detachment of a leaflet*	*Annuloplasty*
Hardy[3]	Transverse	Anterior	No	No
Danielson[11, 12, 18]	Transverse	Anterior	No	Yes
Carpentier[13, 14]	Longitudinal	Anterior	Sliding plasty	Ring suggested
Quaegebeur[15]	Longitudinal	Anterior	Sliding plasty	No
Sebening[16]	None	Anterior	No	No
Current technique	None	Anterior or posterior	Rarely required	Partial annulorrhaphy

than 23 to 25 mm in diameter, equivalent to a valve orifice area of more than 3.6 cm^2.

Although we left the atrialized ventricular chamber unplicated in all patients, echocardiographic serial surveillance revealed no enlargement of the atrialized chamber during the follow-up, and these chambers were mostly the same size as before the operation (Fig. 5).

Discussion

The technique presented here for tricuspid valve repair in Ebstein's anomaly was successfully applied in all 19 cases. These excellent functional results allow extension of the scope of repair, even for cases previously reserved for primary valve replacement. Most tricuspid valve repair techniques rely on the enlarged and mobile anterior leaflet* (Table III), and some patients are not candidates for these procedures because of significant abnormalities of the anterior leaflet, such as inadequate size, attachment of the free edge of the leaflet to the ventricular wall, or failure of delamination.[18] In contrast, our technique allows good repair even in cases where only the posterior leaflet or part of the anterior leaflet can be remodeled to act as a valve-closing structure. The value of this concept is strongly underlined by the stability of the obtained tricuspid competence, now for as long as 9 years of follow-up. To date, there have been no instances of late functional deterioration.

Surgical treatment of Ebstein's anomaly—in particular of its most prominent sequela, tricuspid incompetence—was long disputed because of the immense variety of pathologic features that make the evolution of a standardized concept difficult and also because the scarcity of the defect allowed few surgeons the opportunity to collect sufficiently large experience. Since the initial introduction of repair techniques by Hunter and Lillihei,[6] Lillihei and coworkers,[7] and Hardy and colleagues,[3] repair has not been generally accepted, and valve replacement has been favored as the most straightforward procedure by most surgeons. Danielson's modification of the Hardy technique, applied in more than 300 cases[11, 12, 18] (Danielson GK, personal communication, 1997), used the mobile anterior leaflet as closing mechanism by obliterating the atrialized chamber in a transverse fashion, bringing the body of the anterior leaflet toward the natural anulus and allowing anterior leaflet coaptation with the atrialized septum. Carpentier's repair technique[13, 14] included detachment of the anterior or posterior leaflet at the origin, transection of abnormal chordae if necessary, longitudinal plication of the atrialized chamber, and the reinsertion of detached leaflets to the anatomic anulus above the plicated chamber, also swinging the leaflets toward the posterior septal anulus. Stabilization of this new valve anulus with a prosthetic ring has been proposed.[13, 14]

The crucial questions that must be raised relate to the importance of the atrialized chamber, and most repair techniques include either transverse plication or longitudinal exclusion of the atrialized chamber.[3, 10-15, 18] It was argued that when exposed to right ventricular pressure this chamber might have a similarly negative effect on ventricular energy economics as a left ventricular aneurysm on the left ventricle. Furthermore, stasis within the noncontractile sac might promote clot formation. It was supposed that longitudinal plication, resulting in a "ventriculoplasty," might enhance right ventricular function and obliterate the characteristic rhythm disturbances.[13]

Unlike in most valve repair techniques, the atrialized chamber remains untouched if the tricuspid valve is to be replaced, without negative sequelae.[19, 20] In the only valve repair method with no plication of the atrialized chamber, the base of the anterior leaflet chordal attachment was fixed with a

*References 3, 6, 11, 13, 15, and 17.

single stitch to the muscular portion of the atrialized septum, creating a "monocusp valve."[16] The Sebening technique,[16] where valve competence is achieved with a "single stitch" tying the tip of the main anterior chordae to the atrialized septum, does not include any reefing procedure on the atrialized chamber. Excellent long-term results have been achieved with this technique in more than 60 cases[16] (Sebening F, personal communication, 1997).

In all our cases, no attempt was made to obliterate or to reduce this chamber. To date, we have not seen any indication of further enlargement of the right ventricle, or of any other undesirable effects. This has been substantiated by results of echocardiographic studies during follow-up. In contrast, our impression from earlier experiences with other techniques is that transverse obliteration, at least in the more pronounced cases of types B and C anatomy, may cause high tension both on the Hardy sutures[3] and the tissue, with unknown effects on the already abnormal septum and the left ventricle. Furthermore, hope has been expressed by some of us that incorporation of the atrialized chamber into the contracting right ventricle may be beneficial in allowing sufficient right ventricular filling during diastole and could stimulate the remaining musculature in the atrialized chamber wall toward hypertrophy and even contribution to right ventricular contraction. This, however, must be examined in further observation and more detailed studies.

Reduction of the tricuspid orifice at the level of the true anulus has not created stenosis in any case. This is confirmed by normal inflow velocity in echocardiography and also by absence of any clinical signs of possible stenosis of the tricuspid valve. Care must certainly be taken to avoid the His bundle area by placing the sutures on the septum below the natural anulus, at least in the anteroseptal area. This was not successful in every case, as demonstrated by two instances of complete heart block necessitating pacemaker insertion. The risk of this complication could, however, be avoided with more experience.

An anatomic classification introduced by Carpentier and colleagues[13, 14] appears to be most valuable in surgical decision making regarding the most appropriate procedure. *Type A* refers to cases with mild to moderate displacement of the septal and posterior leaflet origins, preserving a relatively large trabecularized right ventricular cavity and a smaller atrialized chamber. In *type B* this relationship is reversed, with a large atrialized chamber and a smaller contracting ventricle. Common in both types, however, is a large and mobile anterior leaflet. This is restricted in motion in *type C* by adherence to the anterior right ventricular wall of fibrous bands of abnormal chordae. Finally, *type D* describes those cases where the entire right ventricle is lined by broadly adherent fibrous leaflet tissue, giving rise to what is called a *tricuspid sac.*

According to this classification, our patients' conditions were all type A, B, or C. However, there was still vast variability, particularly in the appearance and size of the anterior and posterior leaflets. In the first patient of this series there was no distinction between those two leaflets, and this common leaflet showed a multitude of fissures and clefts. Such clefts were encountered in the anterior leaflet in another three instances, and the posterior leaflet had a substantial size and mobility in six patients. In one instance there was even a fourth leaflet in the anteroseptal commissure. It was decided to close the common orifice to allow the most mobile and substantial part to become the closing mechanism. In type C anatomy, where the anterior leaflet was broadly adhering to the right ventricular free wall, part of this leaflet was freed by careful transection of adherent free wall chordae to achieve a greater mobility of at least this section of the leaflet being chosen to play a part in the newly created valve.

Among this group of patients undergoing valve repair, there was no case with the characteristics of the Carpentier type D anatomy. Recently such a case was encountered, in which the entire right ventricular cavity was lined with fibrous leaflet tissue with broad adhesions to the muscle wall and only a circumscribed, stenotic ostium below the right ventricular outflow tract was visible. In fact, the pathologic features of this case were almost identical to those elegantly described by Dr. Ebstein in his original 1866 case presentation.[1] No attempt was made to repair this defect; the shallow remaining cavity under the fibrous lining was opened by incisions in this fibrous sac starting from the restrictive ostium, eliminating the stenosis, and a valve prosthesis was inserted below the level of the natural anulus.

The technique presented here, which focuses on the individual pathologic features of the tricuspid valve in Ebstein's anomaly, certainly needs further follow-up and experiences in a larger patient group. It is our impression so far that this technique offers good clinical results with a small risk, even with a highly abnormal pathology.

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REFERENCES

1. Ebstein W. Ueber einen sehr seltenen Fall von Insufficienz der Valvula tricuspidalis, bedingt durch eine angeborene hochgradige Missbildung derselben. Arch Anat Physiol Wiss Med 1866;238-55.
2. Keith JD. Prevalence, incidence, and epidemiology. In: Keith JD, Rowe RD, Vlad P, editors. Heart disease in infancy and childhood. 3rd ed. New York: Macmillan Publishing; 1978. p. 3-13.
3. Hardy KL, May IA, Webster CA, Kimbal KG. Ebstein's anomaly: a functional concept and successful definite repair. J Thorac Cardiovasc Surg 1964;48:927-40.
4. Kirklin JW, Barratt-Boyes BG. Ebstein's malformation. In: Kirklin JW, Barratt-Boyes BG, editors. Cardiac surgery. 2nd ed. New York: Churchill Livingston; 1993. p. 1105-30.
5. Watson H. Natural history of Ebstein's anomaly of the tricuspid valve in childhood and adolescence: an international cooperative study of 505 cases. Br Heart J 1974;36:417-27.
6. Hunter SW, Lillehei CW. Ebstein's malformation of the tricuspid valve: study of a case, together with suggestion of a new form of surgical therapy. Dis Chest 1958;33:297-304.
7. Lillehei CW, Kalke BR, Carlson RG. Evolution of corrective surgery for Ebstein's anomaly. Circulation 1967;35(suppl): I111-8.
8. Barnard CN, Schrire V. Surgical correction of Ebstein's malformation with prosthetic tricuspid valve. Surgery 1963; 54:302-8.
9. Charles RG, Barnard CN, Beck W. Tricuspid valve replacement for Ebstein's anomaly: a 19 year review of the first case. Br Heart J 1981;46:578-80.
10. Hardy KL, Roe BB. Ebstein's anomaly: further experiences with definitive repair. J Thorac Cardiovasc Surg 1969;58: 553-9.
11. Danielson GK, Maloney ID, Devloo RAE. Surgical repair of Ebstein's anomaly. Mayo Clin Proc 1979;54:185-92.
12. Danielson GK, Fuster V. Surgical repair of Ebstein's anomaly. Ann Surg 1982;196:499-504.
13. Carpentier A, Chauvaud S, Macé L, Relland J, Mihaileanu S, Marino JP, et al. A new reconstructive operation for Ebstein's anomaly of the tricuspid valve. J Thorac Cardiovasc Surg 1988;96:92-101.
14. Chauvaud SM, Mihaileanu SE, Gaer JAR, Carpentier AC. Surgical treatment of Ebstein's malformation-the "Hôpital Broussais" experience. Cardiol Young 1996;6:4-11.
15. Quaegebeur JM, Sreeram N, Fraser AG, Bogers AJ, Stümper OF, Hess J, et al. Surgery for Ebstein's anomaly: the clinical and echocardiographic evaluation of a new technique. J Am Coll Cardiol 1991;17:722-8.
16. Schmidt-Habelmann, P, Meisner H, Struck E, Sebening F. Results of valvuloplasty for Ebstein's anomaly. Thorac Cardiovasc Surg 1981;29:155-7.
17. Shore DF, Wong P, Paneth M, Buckley MJ. Results of mitral valvuloplasty with a suture plication technique. J Thorac Cardiovasc Surg 1980;79:349-57.
18. Danielson GK, Driscoll DJ, Mair DD, Warnes CA, Olivier WC Jr. Operative treatment of Ebstein's anomaly. J Thorac Cardiovasc Surg 1992;104:1195-202.
19. Sealy WC. The cause of the hemodynamic disturbances in Ebstein's anomaly based on observation at operation. Ann Thorac Surg 1979;27:536-46.
20. Scaly WC. Discussion of Westaby S, Karp RB, Kirklin JW, Waldo AL, Blackstone EH. Surgical treatment in Ebstein's malformation. Ann Thorac Surg 1982;34:394-5.

Discussion

Dr. Guillermo O. Kreutzer *(Buenos Aires, Argentina).* Dr. Hetzer, have you considered performing a bidirectional Glenn shunt beside the tricuspid repair in patients with huge right atrium and poor right ventricular function? We have performed this "one-and-a-half" repair, in addition to tricuspid repair, in eight patients with Ebstein's anomaly and have had excellent immediate and midterm results. We consider this option one more way to improve ventricular function by reducing preload of the right ventricle and right atrium while also diminishing the tendency toward arrhythmia by decreasing the high right atrial pressure.

Dr. Tirone E. David *(Toronto, Ontario, Canada).* Do you refer to type B anatomy, the one with a very small right ventricle and a large atrialized ventricle, or any type of Ebstein's anomaly?

Dr. Kreutzer. No, only for patients with huge right atrium and poor right ventricular function.

Dr. Hetzer. I think this is a valid option. In fact, the first cases of correction of Ebstein's anomaly in the 1950s were such in which a Glenn shunt had been performed to begin with and where the inferior vena cava return was directed toward the right ventricle. I think this may well be an option if you have an extreme case. In our experience we recently saw such type D anatomy and could not do a repair, so we did a valve replacement at the level of the anatomic anulus and just opened the membrane between the atrialized chamber and the trabecularized chamber to alleviate the tricuspid stenosis, which is often present in those cases. But I think it's an option to do a Glenn shunt.

Dr. David. Did you measure the transvalvular gradient by echocardiography after repair?

Dr. Hetzer. Yes, they were all measured. Of inflow velocities on echocardiography, the highest was 1.3 m/sec. Except for three cases, however, the velocity was below 1.0 m/sec in all instances. So although the valve looked quite narrow after repair, there was no functional tricuspid stenosis. I have taken care that after repair there is always an opening at least 2.3 or 2.5 cm in diameter, which gives a valve opening area of at least 3.5 cm^2.

Dr. Dominique R. Metras *(Marseille, France).* I just would like to know if this was a consecutive series, and whether you consider that the tricuspid apparatus sometimes looks terrible, is totally irreparable, and does need a replacement.

Dr. Hetzer. I did not have time to go into further details. Actually, this series has been consecutive since 1994 in 16 cases. That means the three earliest patients were still operated on according to the impression of the individual case. Because we saw that this concept of creating the valve at the level of the anatomic anulus works well, we have used only this technique since 1994. As I mentioned, however, we recently had a case that we considered not repairable, and we did a valve replacement.

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Dr. D. Glenn Pennington *(Winston-Salem, N.C.).* Many of these patients have very large hearts, atria, and right ventricles. I think this is the situation to which one of the other discussants was referring. In this era of volume reduction, have you plicated a right ventricle? Do you routinely reduce the size of the right atrium, for example? And if you don't plicate the atrialized portion, do you end up with a very large heart? Also, how do you deal with Wolff-Parkinson-White syndrome, or have you?

Dr. Hetzer. Of course, there is concern about the size of the heart. I must say, however, that in patients who have huge hearts the hugest part is the right atrium, not the atrialized chamber. I have not encountered a case in which the right atrial chamber was so huge that I thought it necessary to plicate it. On the other hand, from my earlier experience with the Hardy technique and the Danielson technique, I found that if you have larger atrialized chambers and you plicate them in a transverse form according to Hardy or Danielson, you may place an excessive amount of tension on the remainder of the ventricles, both on the right and on the left. It is to this that I attribute some of the negative outcomes.

I think, of course, the right atrium was reduced massively in all cases, and this certainly gives a lot more space to the heart. On the other hand, if you incorporate this atrialized chamber into the functioning right ventricle you may have better diastolic filling, and there may be even a chance for the scarce musculature in the wall of this atrialized chamber to gain function. At least I think there is some hope for that.

There was one case with a definite Wolff-Parkinson-White syndrome in which we had a catheter ablation done after the operation. But we have seen also in some instances that supraventricular tachycardias disappeared after the correction without any specific treatment.

Dr. Norberto G. De Vega *(Malaga, Spain).* I notice that in some cases you repaired the valve by creating two orifices. I am very interested in this concept. I have done some cases in the mitral position to repair the mitral valve, creating a two-orifice valve with very good results. Do you have any experience in mitral repair with this concept? I gather this is Dr. Alfieri's technique.

Dr. Hetzer. Yes. When I read Alfieri's article, I of course thought that this is good concept. We have used this only in one case. But as you know, this has become sort of a routine valve procedure for huge left ventricles, for instance in conjunction with the Batista operation. I think it may well be a valuable option for mitral valve repair as well if it is difficult to use one of the more established types of repair. I completely agree with you.

Dr. João Q. Melo *(Carnaxide, Portugal).* Dr. Hetzer, for the last few months we have operated on patients with Wolff-Parkinson-White syndrome and concomitant significant coronary artery disease. During the bypass grafting procedure, we successfully used radiofrequency catheter ablation under direct operative vision.

Do you believe that in Ebstein's anomaly we can correct the tricuspid valve and the Wolff-Parkinson-White syndrome in the same operation?

Dr. Hetzer. I am about to have a good electrophysiologist join our group, and I think that he can probably handle these special cases in the same way.